<Case Report>Craniosynostosis in Association with Medulloblastoma

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症　　例

Craniosynostosis in Association with Medulloblastoma

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Abstract

A case of a 13-month-old boy with craniosynostosis associated with medulloblastoma was presented. Sagittal suture was prematurely closed, and coronal and lambdoid sutures were widely separated. A large infiltrative tumor in the midcerebellar region was partially removed and treated by irradiation and chemotherapy. A review of the current literature failed to reveal a similar report.

Introduction

Craniosynostosis is known to be related with various pathologies[6]. Although many hypotheses have been proposed, its etiology still remains to be a question. We present a case of premature closure of the sagittal suture associated with medulloblastoma. No similar report was found in the literature.

Case Report

This 13-month-old boy was admitted to our department on October 18, 1990, for clinical investigation of vomiting and slow activity. He was the second child of healthy, unrelated parents. His mother's pregnancy had been normal with no previous history of drug intake. He was delivered by cesarean section after 41 weeks' gestation, and presented at birth an APGAR score of 9, a body weight of 3590 g, and a length of 52 cm. The head circumference was 36.2 cm. His development had been normal until 9 months of age when he started to present some delay in developmental milestones. He was able to sit only at the age of 11 months and still could not stand by himself at the time of presentation. Since one week prior to hospitalization he had had several episodes of vomiting and had been physically debilitated. No family history of neurological disorders was reported.

On examination, the child appeared prostrated and his head was scaphocephalic in shape. The head circumference was within normal range. A skull x-ray revealed a sagittal synostosis associated

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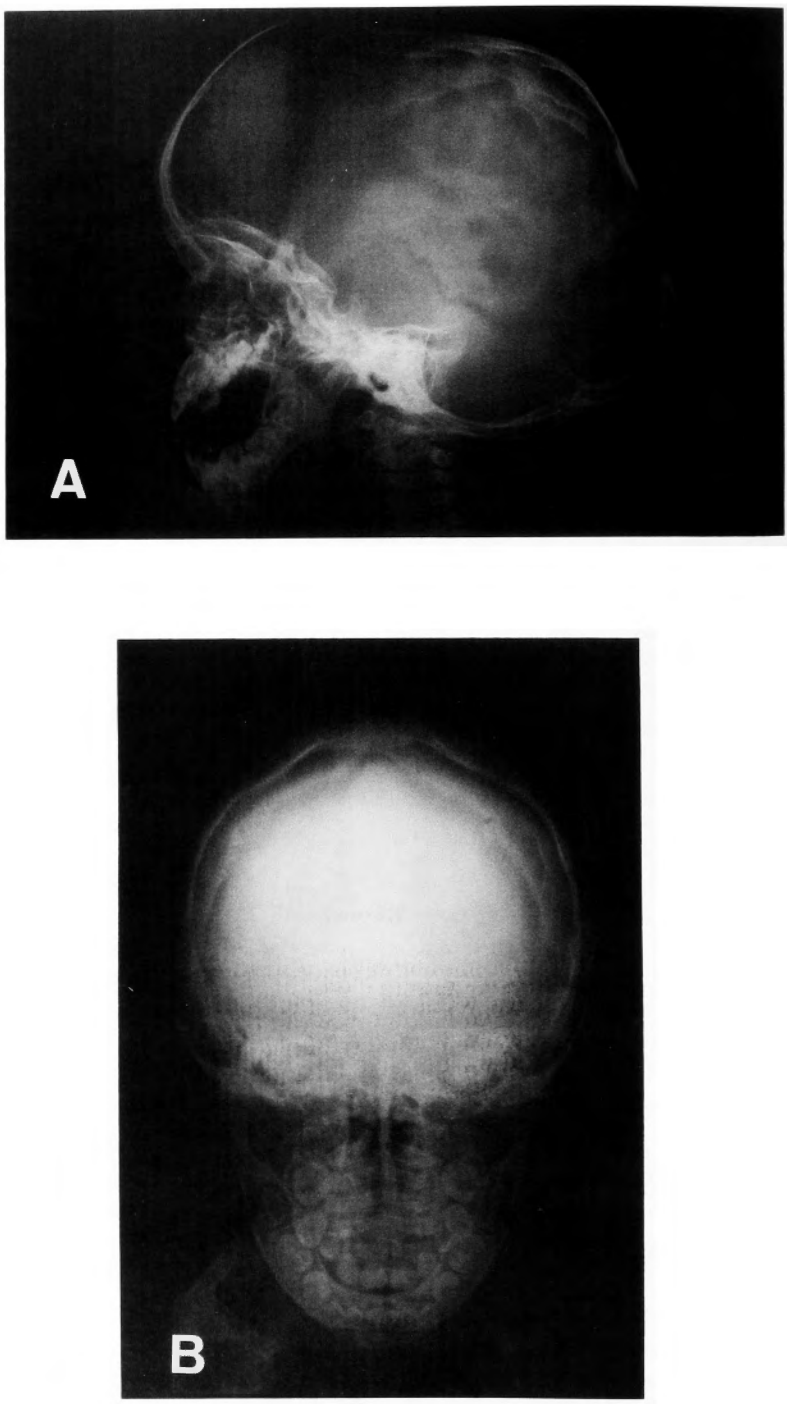

Fig. 1 Plain skull X-ray shows a diastasis of the coronal and lambdoid sutures (A), synostosis of the sagittal suture (B) and digitiforms impressions.

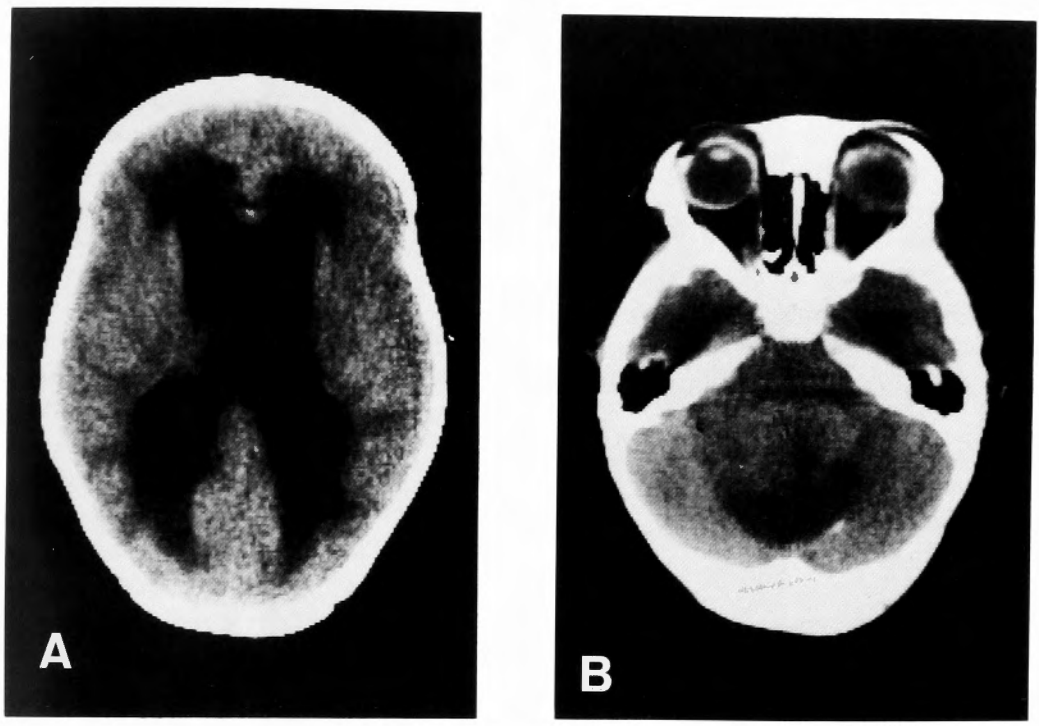

Fig. 2 Non-contrast CT scan shows hydrocephalus (A) due to a cerebellar mass with iso- and hypodensity compared with the brain (B).

with diastasis of coronal and lambdoid sutures (Figure 1). Digitiforms impressions were also present. Laboratory studies including chromosomal analysis were normal. A CT scan showed a huge mass with a cyst in the midcerebellar region, and a moderate hydrocephalus (Figure 2). On MRI the mass was low in intensity on T1 weighted image and enhanced with gadolinium-DTPA infusion (Figure 3). On T2 weighted image and proton density-weighted image, the mass showed high intensity.

A right ventriculo-peritoneal shunt and a suboccipital craniectomy were performed first in order to obtain decompression. About 70% of the tumor was removed in the following two-staged operation. The histological diagnosis was medulloblastoma. A lumbar puncture revealed tumor cells in the liquor. One month after the second tumor removal the patient underwent a single course of intravenous chemotherapy consisting of ACNU (3 mg/kg), vincristine (1 mg/m^2) and cisplatine (80 mg/m^2). Radiation was then initiated and a full brain dose of 23.8 Gy with a boost to the posterior fossa of 19.5 Gy was administered. The whole spine received 21 Gy. A control MRI after irradiation showed a significant decrease in the size of the residual tumor. He was discharged in January, 1991, and is receiving further courses of chemotherapy at the out-patient clinic.

Discussion

The incidence of primary craniosynostosis is supposed to be 3 to 5 per 10,000 live births. The

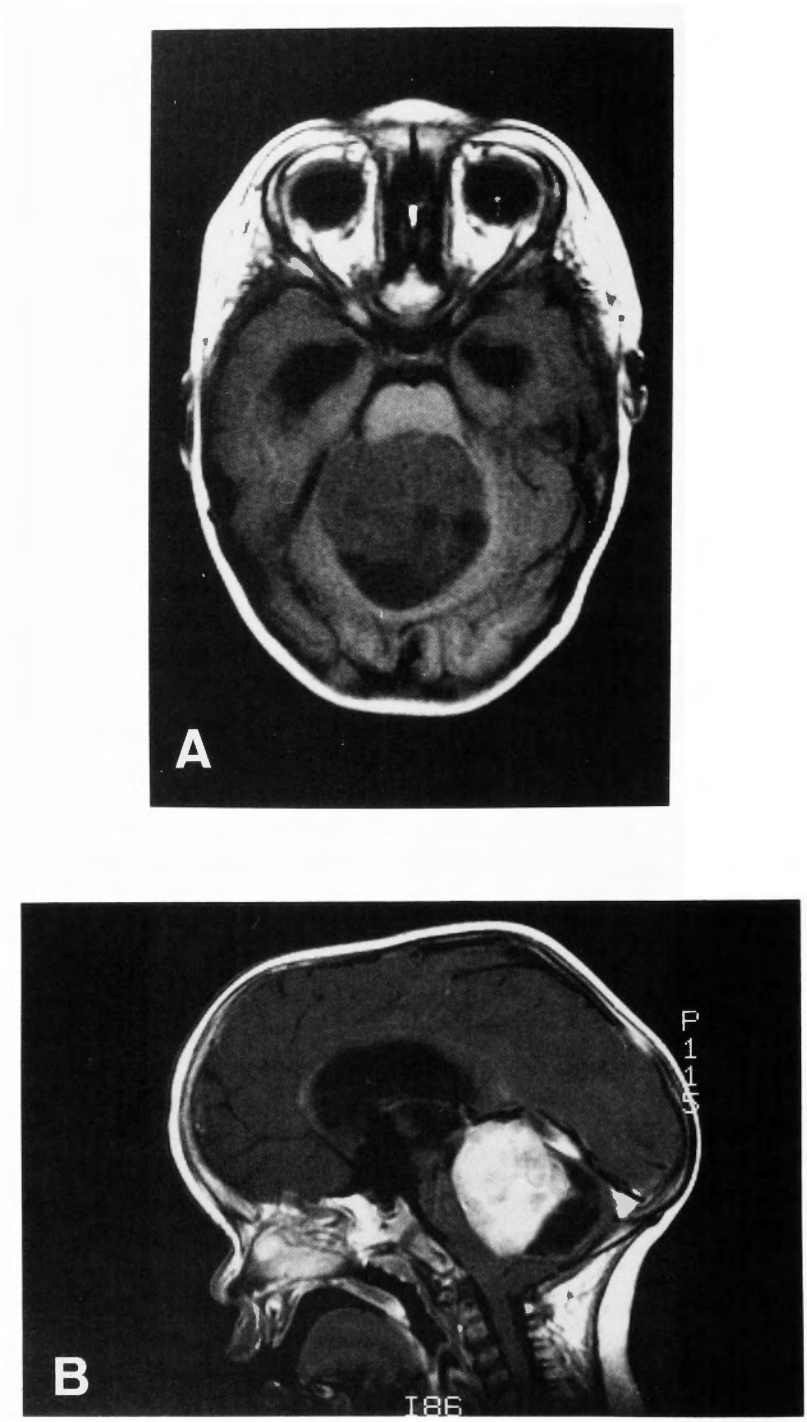

Fig. 3 MRI shows a large midcerebellar mass with low intensity on T1 weighted image (A) and marked enhancement by gadolinium-DTPA (B).

exact incidence, however, is not known. Of equal uncertainty are the pathological mechanisms for premature fusion of sutures[18]. After measuring alterations in the posterior fossa of synostotic patients and after experiments in rats, Moss[14,15] suggested that:

1) The calvaria, dura-mater and cranial base form a single biomechanical entity.

2) A primary malformation of the cranial base produces abnormal forces within its attached dural fiber tracts.

3) This produces craniosynostosis.

Babler and Persing[2,3], from their experimental results on rabbits described that the alteration of the posterior fossa instead of being primary, is secondary to the craniosynostosis. Many papers[1,5,16] supporting or questioning those ideas have been published, but the etiology of the premature closure of sutures still remains to be solved. In the present case a posterior fossa tumor was associated with a sagittal craniosynostosis. To date no similar case has been reported, and most probably it is a coincidence that two different pathologies developed in the same patient. Some authors have already reported craniosynostosis associated with a variety of diseases[4,8,10,11,12,13].

From the diagnostic point of view, the present case shows a somewhat tricky situation. In a child with unfused sutures who harbors a brain tumor with an increased intracranial pressure (ICP), we expect a progressive separation of sutures. Among 19 cases of infra-tentorial tumors which were found during the first 12 months of life studied by Tomita and McLone[19], 10 presented macrocephaly. But, in our patient macrocephaly was absent and the head circumference was within normal limits in spite of an increased ICP due to an obstructive hydrocephalus.

Golabi et al.[8] reviewed 250 children treated for craniosynostosis, and reported that the incidence of hydrocephalus was 4%, and much higher in children with defined craniofacial syndromes than with isolated, simple synostosis. They noted that hydrocephalus develops without either marked head enlargement or signs of increased ICP. Noetzel et al.[17] evaluated 27 children with simple craniosynostosis. None had an evidence of hydrocephalus on CT scans. According to Fishman et al.[7] a communicating hydrocephalus is most common in the craniosynostotic cases. Therefore, clinical manifestations of increased ICP in patients with single craniosynostosis must be carefully investigated, and in case of an obstructive hydrocephalus, a tumoral process should be taken into consideration.

Finally we must discuss the treatment established. Craniectomy presents a good to excellent cosmetic result in more than 90% of patients with sagittal synostosis. Normal mental and motor development is the rule[18]. However, in the present case, an operation for sagittal synostosis is out of indication due to the poor prognosis of the associated medulloblastoma[9]. But, in an infant with an intracranial malignancy who presents with multiple suture stenosis, a corrective surgical procedure may become necessary even in the face of a short life expectancy.

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和文抄録

頭蓋骨早期癒合症に伴った髄芽腫の一例

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頭蓋骨早期癒合症は様々な先天異常や水頭症に合併することが知られているが，われわれの検索した限りでは，脳腫瘍への合併は報告されていない．矢状縫合早期癒合症を伴った髄芽腫の一例を報告する．

症例は13ヵ月の男児で，9ヵ月までは正常の発育を示したが，その後の発育遅延を指摘されていた．入院7日前より嘔吐が出現し，CT 上小脳腫瘍と水頭症を認められ，当科に紹介された．入院時，傾眠傾向を示し，頭囲は 49 cm で拡大はないが舟状頭蓋を示した．頭蓋X線単純撮影では，矢状縫合の早期癒合と冠状，人字縫合の離開を認めた．CT と MRI にて小脳虫部に嚢胞を伴った巨大な腫瘍を認め，水頭症を合併していた．V-P shunt 後，腫瘍の剔出をおこない，組織学的には髄芽腫であった．術後，化学療法および放射線療法を行なった．この症例では，高度の水頭症があるにもかかわらず，頭蓋骨早期癒合があるため頭囲の拡大のないことが特徴的であった．また，腫瘍と頭蓋骨早期癒合の合併の間には因果関係はないものと考えられた．